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WITH REPORT OF THE PATHOLOGICAL EXAMINATION.

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Reprinted from the POST-GRADUATE for March, 1905.

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The group of unusual clinical and pathological conditions connected with the following case is considered by recent German writers to constitute a special form of disease.

On April 14, 1903, the patient, a gentleman aged 54, first presented himself for treatment. He was in good general health with no history of rheumatism, gout or tuberculosis. Syphilis was carefully excluded. For the preceding six months he had noticed progressive loss of vision in his right eye. The refraction was as follows:

O.D. with —6.5 D. sph., with —2.D. cyl., axis 180, vision 6/200.

O.S. with —4.5 D. sph., vision —20/20.

Glasses of the above formula had been worn for 20 years. In the right eye the tension was +1. The optic nerve entrance was white and deeply excavated. The visual field showed glaucomatous limitation with scotoma. He had no pain. There was neither congestion of the conjunctiva nor infiltration of the cornea. The left eye appeared unaffected. The case was diagnosticated as an aberrant form of glaucoma, and eserin in castor oil, followed by water fomentations, were ordered.

The patient was seen again January 11, 1904. He had no

*From the Pathological Laboratory of the New York Post-Graduate Medical School and Hospital.

vision in the right eye. Tension was +1. The optic nerve was very white and the excavation had deepened. At this time he exhibited a *staphyloma-like swelling on the temporal side of the sclera.* Still *no pain.*

At his next visit, on November 18, he stated that two weeks before, while traveling by rail, a valise fell from an overhead rack, inflicting a severe blow on his right temple and eye. This was followed by persistent pain and lachrymation. The pain was intense, requiring morphin at night. There was now deep circumcorneal congestion, chemosis and infiltration of the corneal periphery. The staphylomatous swelling was much larger. Both the eyeball and staphyloma were soft and fluctuating. An unusual depth of the anterior chamber was observed. It was decided to enucleate the eye, and the operation was performed on the following day. The extreme softness of the globe rendered its removal somewhat difficult. A prompt recovery followed, and the patient has since been entirely free from pain. The left eye has shown no signs of the disease.

PATHOLOGICAL REPORT.

As the most important feature in this case is the presence of scleritis, I shall describe only the pathological changes connected therewith and eliminate, so far as possible, those attributable to the coexisting myopia and glaucoma.

Few diseases equally frequent and important as scleritis have been so little studied microscopically. The only cases hitherto examined present such variations in their pathological anatomy that investigation of a new case is essentially research work.

The eye in our case was fixed in 10 per cent. solution of formalin, hardened in alcohol of increasing strengths, and blocked in celloidin. After dividing the eye antero-posteriorly, one-half was sectioned meridianally, the other half equatorially. Sections were cut in series and stained by the following methods: Van Giesen's, hematoxylin and eosin, Weigert's fibrin and elastic tissue stains, and carbol-fuchsin for micro-organisms.

After hardening the following points were noted on gross section: Polar diameter, 28 mm.; equatorial diameter, 26 mm.; protuberance over external rectus muscle; white, circumscribed deposit in the sclera surrounding the cornea; anterior chamber

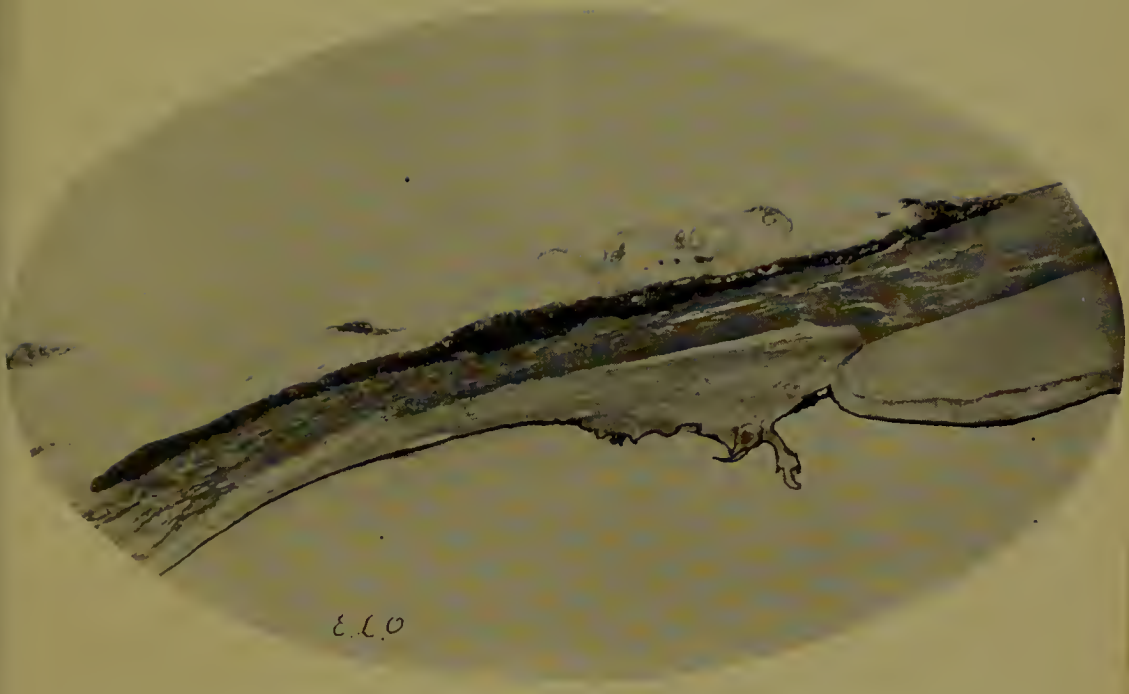

FIG. 1.—Annular scleritis. The principal deposit is in the ciliary body and cornea. Anterior chamber very deep and filled with coagulum. Partial detachment of ciliary body and choroid. B. & L. Obj. 3. Oc.1.

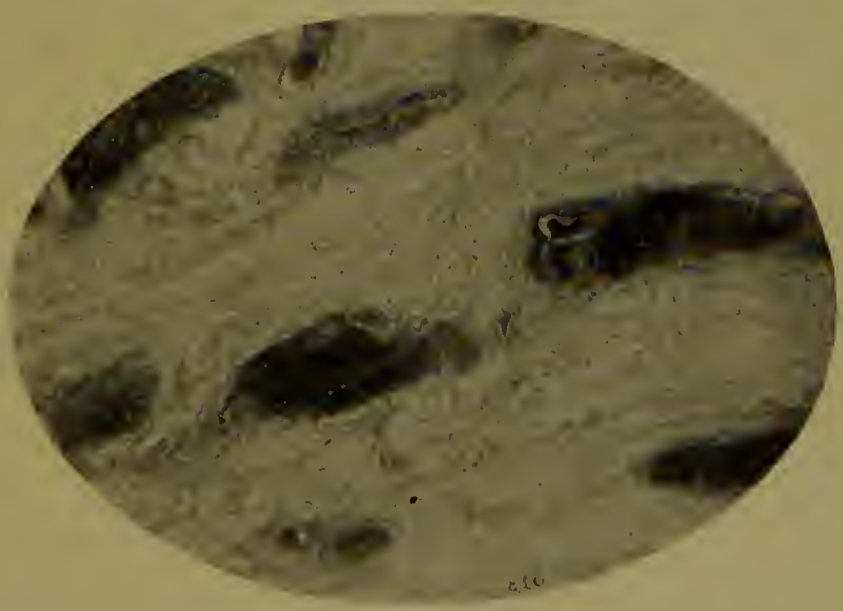

FIG. 2.—Round celled deposits in the loose episcleral tissue. The cells remain in localized, showing no tendency to *infiltrate* the surrounding lymph spaces. B. & L. Obj. 2/3. Oc. 1 1/2.

very deep and filled with white coagulum; white, filmy coagulum in anterior fourth of vitreous; optic nerve deeply excavated.

Microscopical examination shows the anterior regions of the sclera and episclera occupied by a dense cellular deposit which completely encircles and invades the cornea. From the corneal margin it extends backwards about 10 mm. on the temporal and 4 mm. on the nasal side. It is sharply circumscribed and elevated above the surface. On the temporal side are areas where it occupies nearly one-half of the external scleral substance, which it has supplanted (Fig. 1). Isolated nodules, identical in structure to the large deposit, are distributed in the adjacent tissues. Deposits occur also in the subconjunctiva and in the tendon of the external rectus muscle. The scleral deposit is a highly vascularized accumulation of small round cells with no intercellular reticúlum or stroma. The cells are of the lymphoid type with relatively large nuclei. Disintegrating scleral fibers are scattered throughout the mass (Fig. 3). The vascularity of the deposits renders Shirmer's comparison to granulation tissue more applicable than that of Uhtoff to trachoma follicles. Neither polynulcear leucocytes, endothelial nor giant cells are found. The microscopic examination for nicro-organisms was negative. The blood vessels are enormously distended with blood; their walls, particularly the veins, are packed with round cells. The deposit develops along the anterior ciliary system of blood vessels. In the overlying subconjunctival tissue the arteries show hyaline degeneration, obliterating endarteritis and endothelial proliferation. Uhtoff describes vessels containing clusters of red blood corpuscles surrounded by a homogeneous coagulum, from which he concludes that great circulatory disturbance had existed. A similar appearance is observed in this case.

Schlodtmann describes the deposit as a *cellular infiltration* between the scleral fibers. This method of development is not strictly applicable to the case under consideration. The cells once deposited show little tendency to wander or *infiltrate* the neighboring lymph spaces. Fig. 2 shows the development of isolated nodules around blood vessels in the loose episcleral tissue. Here the cells remain localized, showing no tendency to wander into the surrounding dilated lymph spaces.

The invasion of a new area is characterized by dilatation of

the existing blood vessels. Apparently the cells first appear in the walls of the vessels, preferably the veins. From the focus thus established the surrounding connective tissue is replaced by round cells (Fig. 4.) It is assumed that the cells are of connective tissue origin and result from proliferation of the fixed connective tissue of the parts.

The subconjunctival lymph spaces are greatly dilated. Cellular deposits are especially numerous around blood vessels situated immediately beneath the conjunctival epithelium (Fig. 5). This peculiarity was observed in Shirmer's case.

In the case under examination the pathological changes observed in nerves distributed to the affected region are of the highest interest, strongly suggesting that *this case of scleritis may be a reflex or trophic disturbance.* In the scleral deposits and subconjunctival tissue, the nerves are in an advanced stage of proliferative interstitial neuritis. There is enormous thickening of the nerve sheath and hyperplasia of neuroglia tissue. Nerves are found in which axis cylinders and medullary sheaths have entirely disappeared; being replaced by fibrous tissue (Fig. 6). The *cornea* is invaded first in its external layers. The epithelium is edematous and its anatomical arrangement disturbed. Bowman's membrane is split and indistinguishable from other disintegrating corneal fibres (Fig. 7).

The *iris* and *ciliary body* are moderately atrophic and edematous. It is probable that the atrophy represents iridocyclitis of the low grade common to chronic glaucoma, while the edema is of more recent origin, resulting from obstructed ciliary circulation. Round cells are not observed in the iris; they are, however, beginning to appear in and around the walls of those vessels in the ciliary body lying nearest the sclera.

The anterior chamber is very deep and filled with a slightly fibrinous coagulum containing a few red blood corpuscles. The condition at the filtration angle is a curious feature of the case. The spaces of Fontana contain considerable pigment. They are now open and would undoubtedly allow free passage to normal aqueous. It is, however, apparent that they were formerly obstructed by adhesion of the periphery of the iris to the cornea. This area of old attachment can be traced by the ruptured adhesions and attenuated iris tissue still adherent

FIG. 3.—Annular scleritis. At this point the external third of the scleral substance has been supplanted by the deposit. Fragments of disintegrating scleral fibers are scattered through the mass. B. & L. Obj. 1/4. Oc. 1.

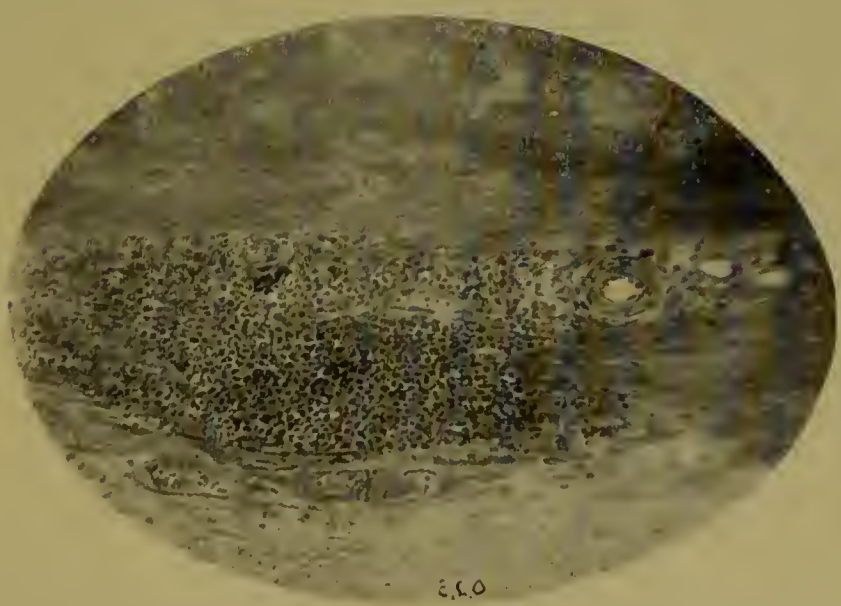

FIG. 4.—Posterior edge of the scleral deposit. The right hand side of picture shows area of new invasion. The cells appear first in and around the walls of the blood vessels. B. & L. Obj 1/4. Oc. 1 1/2.

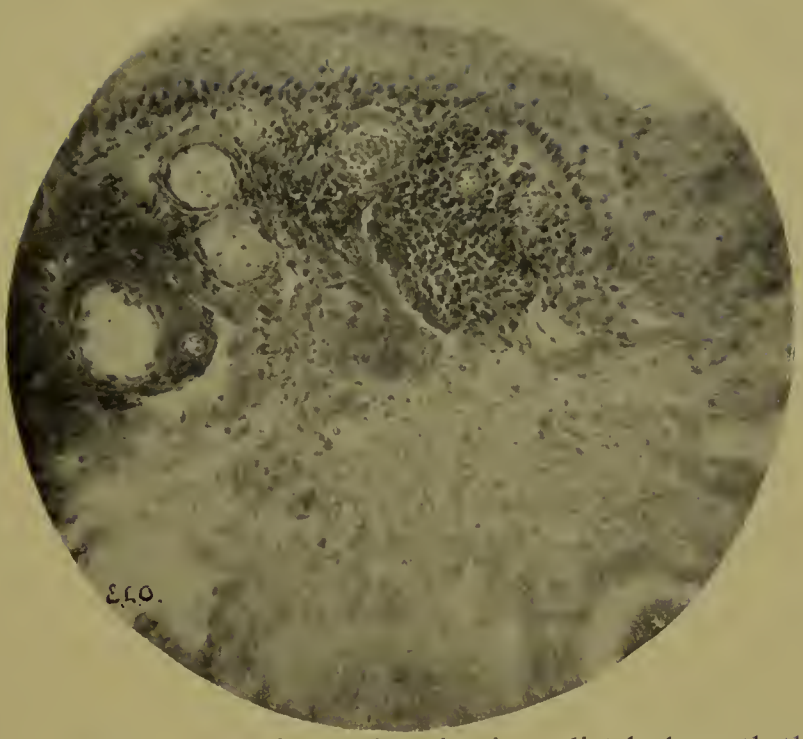

FIG. 5.—Deposits in the subconjunctiva immediately beneath the epithelium. They are particularly numerous in this region. B. & L. Obj. 1/4. Oc. 1 1/2.

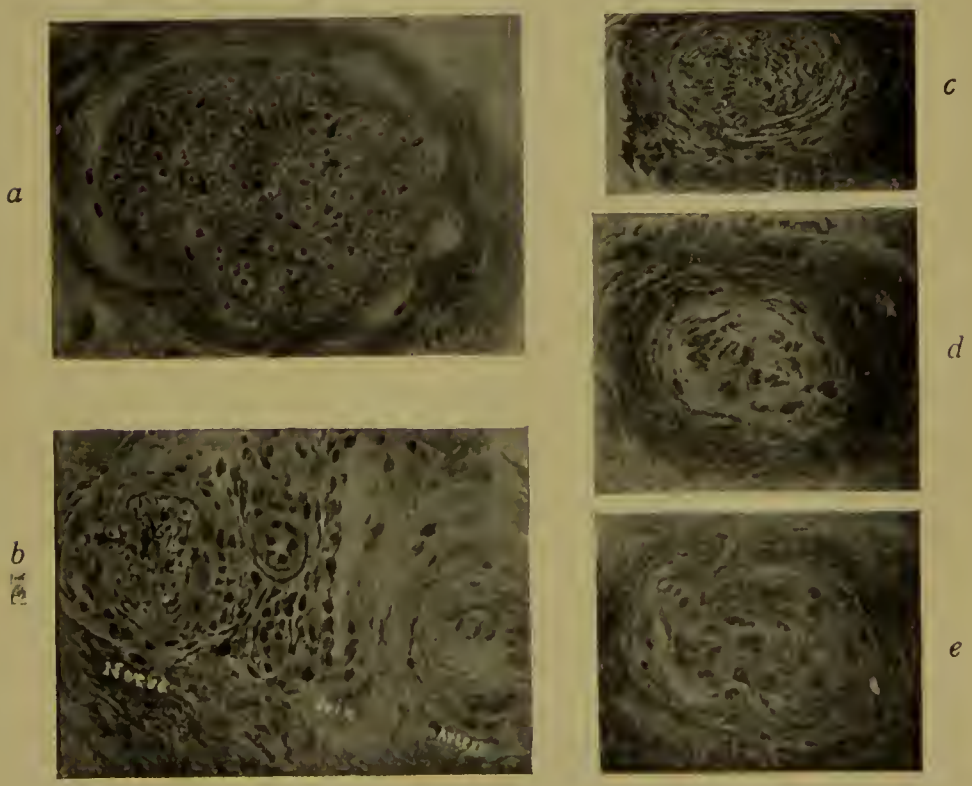

FIG. 6.—Nerves in Annular Sclcritis. *a*, normal ciliary nerve; *b*, *c*, *d* and *e*, peripheral branches of ophthalmic division of fifth nerve in various stages of sclerosis; d and e are entirely converted into fibrous tissue.

to the *ligamentum pectinatum irides*. The iritis being of a low grade, the peripheral adhesions readily yielded to the backward drag of the ciliary body and iris. This displacement of the iris and ciliary body cannot be attributed to the usual cause, namely, the organization and shrinking of inflammatory exudates. The only apparent explanation is pressure in the anterior chamber from the large transudate resulting from an obstructed anterior ciliary circulation.

Another remarkable feature is a large deposit in the tendon of the external rectus muscle. At its point of attachment to the globe, the tendon, tendon sheaths and surrounding sclera are largely replaced by the characteristic deposits (Fig. 8). Small scanty deposits are found at the junction of the externus with its tendon. They are strictly confined to the interstitial connective tissue, the muscle fibers not being affected. The sheaths and surrounding tissues are greatly distended with a serous effusion. The protuberance thus formed in connection with an underlying ectasis of the sclera, constituted the staphyloma observed clinically. At this point is a complete rupture of the sclera, extending from the anterior margin of the tendon of the externus, backwards and inwards to the interior of the eye. Along the line of rupture the scleral fibers show marked degenerative changes, while their interspaces contain coagulated fluid, free pigment and pigmented cells. This communication between the interior of the eye and the external protuberance explains the sudden appearance of extreme flacidity observed in both, as is described in the clinical history. The low tension cannot be attributed to phthisis bulbi or lack of aqueous, the ciliary body being only slightly atrophic. This fistulous opening was probably produced by the following conditions: high intraocular pressure; extensive destruction, weakening and ectasis of the sclera; continuous drag of the external rectus muscle on its diseased tendinous attachment; violent blow on the eye and temple.

It cannot be asserted that new connective tissue has formed from the cellular deposits, yet the changes in the periphery of the densest areas indicate that such a process has commenced. It is probable that our case exhibits an early stage of scleritis, the unique occurrence of a fistulous opening leading to early enucleation.

There is widely disseminated atrophic degeneration of the *choroid*. Opposite the scleral deposit the sclera, choroid and retina are intimately united by plastic adhesions. The remaining choroid is widely separated from the sclera by effusion into the suprachoroidal space of a fluid similar to that found in the anterior chamber. No round celled deposits are found in or around the walls of the venae vorticosae. These vessels show no evidence of obstruction.

The *retina* is irregularly degenerated. Peripheral cystoid degeneration (Iwanhoff's edema) exists opposite to the scleral deposit.

With our present limited knowledge of scleritis a scientific classification of the different varieties is impracticable. One form has been described by Schlodtmann, in Fuchs' clinic, under the name of " Sulziger Infiltration der Sclera." Friedland and Uhtoff subsequently reported cases which they placed in this class. Parsons accepts the classification but suggests " Annular Scleritis " as a preferable designation.* Although the cases thus far examined show great morphological variations, they resemble each other clinically. It is a disease of advanced life. The nodules coalesce and completely encircle the cornea which also is invaded. It is associated with slowly progressive uveal disease, usually terminating in loss of vision. These points serve to differentiate it from the nodular recurring scleritis of young adults affected with syphilis or rheumatism.

As the case here reported resembles in important points those described by the above German writers, I shall venture to place it in the same category.

Uhtoff believes that this form of scleritis is a special disease. This view is sustained by the complexity of symptoms in our case. The occurrence, in the same eye, of myopia, glaucoma with persistent high tension, extensive scleritis with ectasis, terminating in blindness previous to the occurrence of pain or external signs of inflammation, is very unusual.

The etiology of scleritis unassociated with constitutional taint is very obscure. Uhtoff justly regards the vascular changes and consequent obstruction as accountable for many of the phenomena observed. It appears certain that the edema of the

* Parsons rejects as spurious the only case examined microscopically by Schlodtmann.

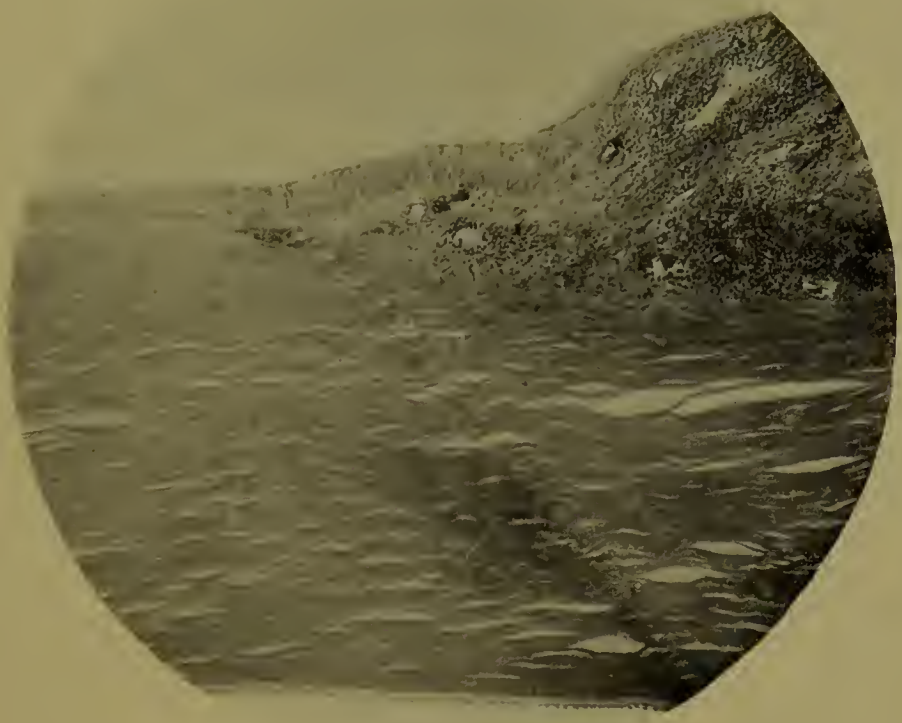

FIG. 7.—Invasion of the cornea by round celled deposit in annular scleritis. B. & L. Obj. 2/3. Oc. 3/4.

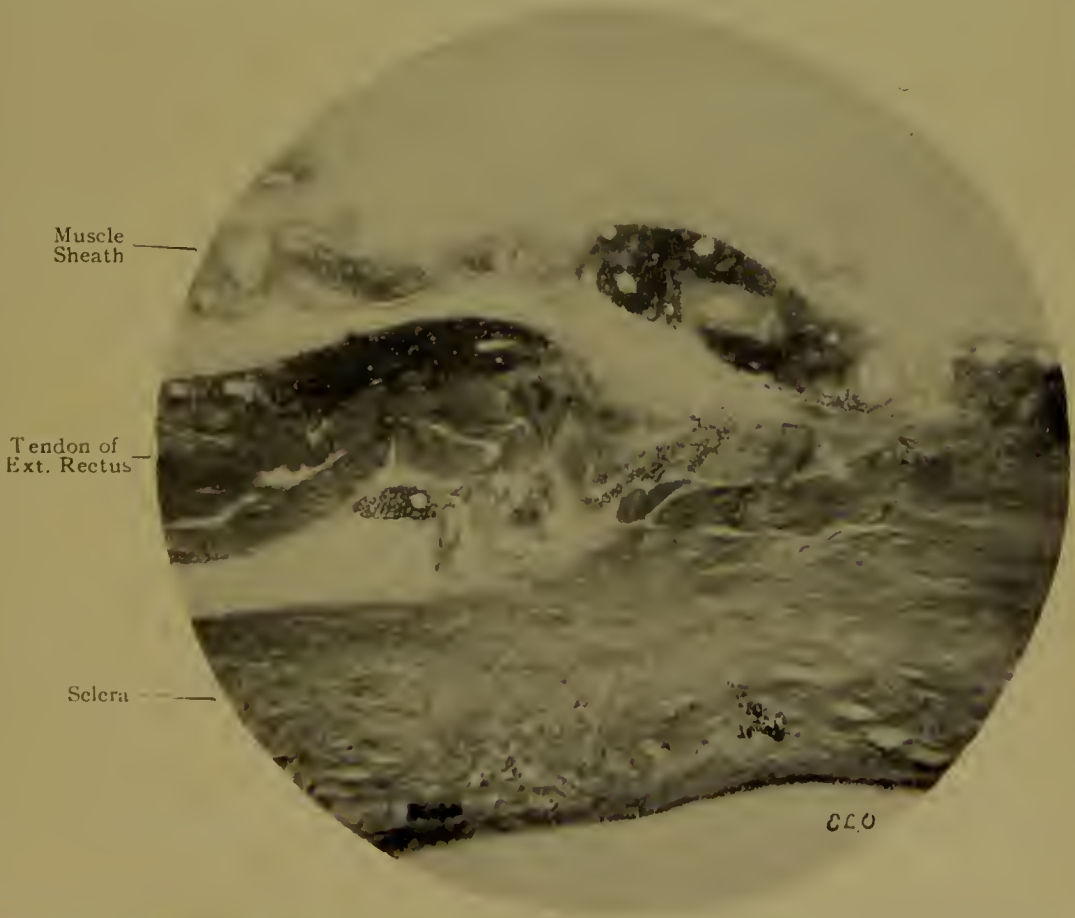

FIG. 8.—The sclera is ruptured near this point. Large deposits are

iris and ciliary body, and the effusion into the anterior chamber, which we have described, must be attributed to obstruction of circulation in the anterior ciliary vessels. The cause of the vascular degeneration, however, remains unexplained.

Among the puzzling features of this disease, are its limitation to the anterior half of the globe, its usual origin in the episclera, the frequent absence of pain until the uvea is deeply involved, and the absence of pathognomonic tissue changes. The theory I have here suggested, that some cases of scleritis may be trophic, dependent on nerve or ganglionic degeneration, at least offers a satisfactory solution to the enigma. In our case this theory is supported by the fact that the principal seat of the scleritis is the area of distribution of the ophthalmic branches of the fifth nerve, which are the only nerves found to be diseased. Furthermore, the ophthalmic nerve has anatomical connections with the abducens, which supplies the external rectus, whose tendon is affected, and, what is more significant, it communicates with the cavernous plexus of the sympathetic, which distributes nerves to the anterior ciliary vessels, around the branches of which the cellular deposits occurred.

That the nerve degeneration is not a result of the surrounding scleritis but preceded it, is indicated by the following facts: (1) The scleral changes belong to the early stage of a formative inflammation; (2) in the nerves formative inflammation is very far advanced; (3) the scleritis proceeded to staphyloma formation without pain—the late pain, which followed the blow, probably was due to detachment of the ciliary body by effusion; (4) degenerated nerves were found in the loose subconjunctival tissue apart from the cellular deposits.

Reflex or trophic eye lesions, which may include sympathetic ophthalmitis, scleritis, herpes, some forms of keratitis and possibly other diseases, have not yet been placed upon a satisfactory scientific basis. Nevertheless, many investigators still adhere to the faith of the fathers of Ophthalmology who saw in the reflex theory of disease a satisfactory explanation for many clinical phenomena. Theobald, in a recent paper supporting the reflex origin of sympathetic ophthalmitis, says: "Intense and long-continued irritation of sensory nerves may cause inflammation in ganglion cells with which these nerves are in relation, and these in turn may bring about inflammatory changes."

This view is founded on recent scientific investigations. He says also, " I have looked on . . . trophic nerve affections . . as due to disease of one or more of the ganglia in relation with the fifth nerve . . . or of the central ganglia themselves."

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